In 2000, a 46-year-old male with history of recurrent airway infections developed episodes of abdominal pain, arthritis and palpable skin purpura without overt renal involvement. He had serum IgG 377 mg/dl, IgM 20 mg/dl, normal IgA (increased during relapses of cutaneous vasculitis) and low CD19+ B-cells (See Tab 2). Causes of secondary humoral immunodeficiency have been ruled out (See Tab 1). Poor response to vaccination was observed. Skin biopsy showed the presence of leukocytoclastic vasculitis with endothelial deposits of IgA. A diagnosis of Henoch-Schoenlein purpura (HSP) concurrent with Common Variable immunodeficiency (CVID) was reached, and he has been treated over the years for multiple relapses with steroids, intravenous and oral cyclophosphamide (CYC), methotrexate and replacement with intravenous immunoglobulins.

During the last HSP relapse, he also developed dyspnea and hemoptisis. Chest CT showed bilateral patchy alveolar infiltrates suggesting pulmonary haemorrhage and capillaritis, confirmed by brochoalveolar lavage. Based on most recent experiences on the use of B-cell depleting agents, such as rituximab, in vasculitis and in different autoimmune manifestations, even in the context of CVID, we chose to use Rituximab (RTX) after failure of steroids. He received two 800 mg doses of RTX at 2-week intervals and a single i.v. 500 mg dose of CYC. He was discharged after a rapid regression of cutaneous, abdominal and respiratory symptoms, confirmed by clearing of infiltrates at chest-CT and normalization of laboratory findings.

He is now doing well and the re-treatment with RTX is planned at B-cell reconstitution. Diffuse alveolar hemorrhage is a life-threatening manifestation of pulmonary vasculitis. Pulmonary involvement in HSP is rarely observed. We describe an adult patient affected by CVID and recurrent HSP, successfully treated with RTX for occurrence of alveolar haemorrhage. Further experience is warranted to assess the efficacy of RTX for pulmonary involvement in vasculitis. Additionally, the use of RTX or other B-cell targeted treatments in difficult-to-treat forms of Henoch-Schoenlein purpura deserves more clinical experience.